356 Vitamin K deficiency is present in cystic fibrosis patients from infancy

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Cystic fibrosis (CF) is an inflammatory and destructive disease with differentiated clinical expression. Conducted studies revealed the presence of vitamin K deficiencies. However, available results in infants and young children are limited. Therefore, the aim of the present study has been to assess vitamin K status in young CF patients.

Material and Methods: Twelve CF infants aged up to 12 months were introduced into the study. Uncarboxylated prothrombin concentrations (Prothrombin Induced by Vitamin K Absence – PIVKA II) as vitamin K deficiency index were measured (ELISA) in all subjects. In addition, nutritional status (standardized body weight), exocrine pancreatic function (fecal elastase-1 concentrations), expression of bronchopulmonary disease and vitamin K dosage were also assessed.

Results: All subjects were pancreatic insufficient, pulmonary manifestation was differentiated. In a significant percentage of CF patients body weight deficit was present at diagnosis. Eight children received vitamin K preparations in a dose 0.025–0.4 mg/day. In five children receiving supplementation and two non-supplemented subjects PIVKA II was detectable (range 3.1 to 97 ng/mL). No significant correlation between PIVKA II levels and values of classic ($r = -0.468$) and conductometric ($r = -0.454$) sweat test, standardized body weight ($r = 0.186$), fecal elastase-1 concentrations ($r = 0.116$), vitamin K supplementation dose ($r = 0.334$) and the expression of bronchopulmonary disease ($r = -0.230$) was stated.

Conclusions: Vitamin K deficiency seems to be present in cystic fibrosis patients from infancy. No correlation between PIVKA II and parameters assessing the intensity of the disease has been stated.

357* Predictors of vitamin K status in children and young adults with cystic fibrosis

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Predictors of vitamin K status were examined in 8- to 25-yr old subjects with cystic fibrosis (CF) and pancreatic insufficiency (PI). Growth status, genotype, pulmonary function, serum vitamin D, dietary intake, vitamin K supplemental intake, and serum undercarboxylated osteocalcin as % total osteocalcin (%ucOC) were assessed. Sufficient vitamin K status was defined as %ucOC < 20%. Subjects were divided into 3 vitamin K supplementation groups: <150 ug/d (low; multivitamins/no supplement), 150–999 ug/d (mid; CF-specific vitamin preparations), and ⩾1000 ug/d (high; mephyton). Vitamin K supplementation (median intake 300 μg/day) and %ucOC (34±18%) were negatively associated ($r = -0.55$, $P < 0.001$). Only subjects in the high supplemental intake group achieved a sufficient vitamin K status (22±16) compared to the mid (38±16) and low (45±14) groups (both $P < 0.001$, high vs. mid and high vs. low). Age, gender, vitamin K supplementation group, and vitamin D were significant predictors of vitamin K status, together explaining 43% of the variance in vitamin K status. By gender, vitamin K supplementation group for both males and females, in addition to vitamin D and a trend for age in males were significant predictors of vitamin K status together explained 31 and 54% of the variance in vitamin K status in males and females, respectively. In conclusion, only vitamin K supplementation dose predicts vitamin K status for both males and females with CF and PI, thus highlighting the importance of routine high dose vitamin K supplementation for this population.

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358 Oxidative stress in stable CF patients: do we need higher antioxidant plasma levels?

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Background: Increased oxidative stress is well known in Cystic Fibrosis (CF) patients. Studies still address the question if CF patients need higher antioxidant plasma concentrations than healthy subjects.

Objective: To assess if "normal" values of plasma antioxidants counterbalance adequately oxidative stress in stable CF patients with or w/o pancreatic insufficiency (p.i).

70 patients were recruited during a routine visit in our CF Regional Center (32M/38F, median age 7.8, 36/70 with p.i.) whose 91% with vitamin supplementation. Red-ox status as: plasma or erythrocytes antioxidants: vit. E, A, C, selenium (Se), reduced (GSH) and oxidised (GSSG) glutathione, GSHPx; lipid oxidation markers: chromolipids (MDA and HNE aldehydes).

Results: The majority of patients (>87%) showed normal concentrations of vit E, A, C and Se (local reference range). Similarly GSH and GSHPx erythrocyte levels were normal (>74%). Actually normal chromolipid levels were found in only 2–6% and 94% of patients showed low GSH/GSSG ratio. CF patients with p.i. showed significantly higher HNE- and MDA-chromolipid levels, despite the absence of steatorrhea and no significant differences in vitamin concentrations. Independently from other viariables, significant correlations were found between age and vitamin E and C concentrations (inverse), and MDA, HNE chromolipids (direct) and between antioxidants and MDA or HNE chromolipids (inverse).

Conclusions: "Normal" antioxidant plasma levels do not provide adequate control on oxidative stress markers that are directly correlated with aging, pancreatic insufficiency and indirectly correlated with plasma levels of antioxidant vitamins.

359 Iron status of paediatric cystic fibrosis patients

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When reviewing annual review results we found that of our CF patients had normal Hb but that majority had a low RBC mean cell volume (MCV) a feature which is seen in iron deficiency. We therefore started to measure serum ferritin as a measure of iron status at annual review and assessing the iron intake of patients from their 3 day food diaries.

Method: Latest annual review FBC and ferritin results and analysed food diaries (where available) were studied. Information on whether the child was on iron supplements was obtained from notes.

Results: Latest annual reviews of 34 CF patients (aged 1 to 15 years) were looked at. 32/34 were pancreatic insufficient. FBC available for all, serum ferritin for 24 and analysed completed food diaries for 15. 4 patients were already on iron supplements (started previously for low iron intake). Mean Hb was 13.4 g/dl, mean MCV 79.5 fl. MCV for those not on iron supplements were lower (79.4) than those on iron supplements (80.3). Ferritin levels for patients not on iron supplements (21/24) were low (mean 26 ng/ml) compared with those on iron supplements (mean 145 ng/ml). Dietary iron intake was above recommended in majority (12/15). 2 out of the 3 patients with low dietary iron intake were already on iron supplements. 7 further patients were started on iron supplements following their annual review as their ferritin was low (mean 21.3 ng/ml).

Conclusion: Serum ferritin levels of our paediatric CF patients which are not on iron supplements are low despite adequate iron intake suggesting a possible problem with iron absorption. Iron supplementation results in a rise in ferritin levels but is this of any clinical significance?